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A CASE OF RAPID AND WIDESPREAD MUSCULAR WASTING WITHOUT DISEASE OF THE SPINAL CORD.

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THE patient whose history is the subject of this paper entered the Massachusetts General Hospital in May, 1879, under the care of Dr. S. L. Abbot, who very kindly allowed me to see and investigate the case, and subsequently to examine the spinal cord, and now permits me to make full use of his clinical notes. My thanks are due both to him and to his then clinical assistant, Dr. W. P. Gannett.

The essential features of the patient's last illness were as follows: It was an acute febrile attack, ending fatally in about one month, and characterized by severe pain, diminution of sensibility, rapid muscular wasting, and diminution of electrical irritability in all four extremities, mainly confined to the parts below the elbows and the knees, also by alteration of the mental condition, and delirium.

After death, spots of softening were found in the great ganglia of the brain and in the centrum semiovale. The

spinal cord was essentially healthy. The peripheral nerves were not examined, but the inference is drawn that the case was one of disseminated neuritis.

The following are the details of the case:

Margaret C., married, 50 years old, was admitted to the Massachusetts General Hospital May 22, 1879, and gave the following history:

She had always been well, as she thought, until three weeks before entrance. At that time she became chilly and feverish, and supposed herself to have taken cold. She was attacked at the same time with severe pain, which began in the toes of both feet, but extended over the entire body. The use of the legs became progressively impaired, and the hands also grew so weak that in the course of two weeks she became unable to hold anything in her grasp. She had been obliged to keep her bed from the first of the attack, and her sleep had been much disturbed by pain. No nourishment could be retained except milk.

On examination there was found to be no swelling of the joints; a good deal of general tenderness on pressure, not sharply localized; motion at the wrist and ankle and phalangeal joints was found to be greatly impaired, and the cutaneous sensibility of the skin over the feet and ankle joints diminished. Auscultation of the heart revealed nothing abnormal. No reflex movement could be excited by tickling the soles of the feet. The temperature was 100.1° (F.); pulse, 110; respiration, 30. Milk was ordered in small quantities, and salicyl. soda, grs. x, every hour, p. r. n.

The subsequent history of the case is as follows:

May 23d.—Much pain during night. Was given Dover's powder, grs. x. The catamenia appeared during the night.

May 24th.—Was somewhat delirious and quite restless through the night, and is still slightly so. Complains of pain and burning in the feet, but they are not swollen or tender.

May 25th.—Delirious during night; nearly free from pain and delirium this morning. Tongue dry in centre, furred on both sides.

May 26th to 31st.—No marked change.

The report of the examination of the urine (Prof. E. S, Wood) is as follows:

Urine.—Acid; yellow; sp. grav. 1017; urophain slightly diminished; indican and urea normal; uric acid in excess.

Earthy phosphates normal. Alkaline phosphates slightly increased. A slight trace of albumen. Considerable sediment containing numerous hyaline and granular casts, considerable uric acid, excess of mucus, a little blood, clumps of pus, much bladder and vaginal epithelium. Some of the casts have highly refracting granules on them.

May 31st to June 2d.—Complains of no pain except in feet. No tenderness or other abnormal sign about vertebral column. A subcutaneous injection of pilocarpine (gr. ⅓) yesterday caused a profuse sweat.

Her present condition is as follows:

All movements of the arms at the shoulder and elbow, and of the legs at the thigh and knee, are possible, though but slowly and feebly performed. There is no voluntary or reflex movement of the fingers and hands, nor of the feet and toes. The movements of the head are apparently free.

There is well-marked loss of sensibility of the skin of all four extremities, especially of the arms below the elbows, and of the legs below the knees, the intensity of the anæsthesia increasing the nearer the feet are approached.

Within these areas neither contact of the finger nor moderately strong applications of electricity excite any sensation. Applied to other parts of the arms or legs, such currents cause manifestations of pain.

The arms are usually, though not invariably, somewhat flexed, and the biceps rigid.

Under these circumstances the biceps can be excited to contraction by sudden, passive stretching.

The results of the electrical examination at this date were as follows:

Left arm; faradic current; the reaction of the ulnar and musculo-spiral nerves, and of the muscles supplied by them, is preserved but greatly diminished. The biceps and triceps react better, though hardly as well as normal.

The reaction of the median nerve, and of all the muscles supplied by it, as well as that of the interosseous muscles, is entirely wanting.

Galvanic current. The typical degenerative reaction is nowhere present. Strong currents, however, elicit feeble contractions

from most or all of the muscles, if applied directly ; not, however, through the medium of their nerves, except in the case of the median. However excited, the contractions are quick and short in character.

The condition of the right arm is essentially the same with that of the left.

Left (and right) *leg ; faradic current.* The reaction of the quadriceps ext. cruris group and of the hamstring muscles is preserved, though only the feeblest contractions can be excited.

When applied to the peroneal nerve even the strongest currents fail to excite any contraction in the corresponding muscles.

A strong *galvanic* current, on the other hand, applied to the same nerve, excites marked though feeble contractions in all the muscles supplied by it. These contractions appear and disappear more slowly than normal.

Strong *galvanic* currents applied directly also excite contractions in these same muscles. These contractions likewise are slower than normal, and if repeatedly excited their intensity becomes rapidly less (reaction of exhaustibility).

The muscles of the back and of the neck react apparently well.

All the muscles of the body, but especially those which have lost their faradic reaction, are extremely feeble. The interosseous muscles, as well as the long flexors of the fingers, are noticeably atrophied.

All the reflexes (except that of the biceps above alluded to), including also the reflex of the abdominal muscles, and the conjunctival reflex, are wanting. The corners of the mouth are drawn down, giving the face a haggard expression ; but this may be partly due to the fact that the patient is drowsy and but semi-conscious.

The fundus of the right eye, examined with the ophthalmoscope, shows no abnormal appearance.

Passive movements of the arms and legs cause decided expressions of pain.

The patient lies groaning without interruption, though she can be aroused by the sound of her name, etc., when her face often breaks into a senseless smile.

The pupils are equal and about normal in size ; they respond slightly, though promptly enough to light.

June 4th. Condition the same. Patient lies in a stupefied condition, but can be aroused without much difficulty, and says she has no pain.

June 6th. Since the last report the patient has failed very rapidly. Cannot now be aroused. Swallows a little brandy and water with much choking. Passes urine involuntarily, as she has for several days.

The patient sank rapidly during the following night, and died quietly at 4.15 A.M. of the 7th, the temperature having risen through the past two days, reaching 107° an hour before death.

The temp. (axillary) chart of the greater part of the sickness was as follows :

Date.	Temp. (F.).		Pulse.		Resp.	
	A.M.	P.M.	A.M.	P.M.	A.M.	P.M.
May 22d	——	100.4°	——	110	—	30
" 23d	100°	99°	110	100	21	24
" 24th	98.7°	98.7°	110	102	20	25
" 25th	98.6°	99°	100	108	20	25
" 26th	98°	104°	106	104	20	30
" 30th	100°	102°	110	140	17	30
" 31st	100°	102°	108	120	30	34
June 2d	101°	102.4°	110	128	28	32
	Noon.	Midnight.	Noon.	Midnight.	Noon.	Midnight.
" 6th	105.2°	106.4°	110	140	34	39
	3 A.M					
" 7th	107°	Death at 4.15 A.M.				

The treatment employed (salicylic acid at first, subsequently brom. pot. and other palliatives, moderate alcoholic stimulation, milk) had the effect of keeping excitement, pain and fever down to a moderate point, and in view of the acute softening in the brain, discovered *post mortem*, it is improbable that death was directly hastened by any of these conditions.

At the autopsy, which was made by Dr. R. H. Fitz, the organs of the chest and abdomen were found to present no abnormal appearance. The kidneys were not further examined, but the condition of the urine, together with the fact that the arteries throughout the spinal cord were subsequently found notably thickened, suggest that, nevertheless, some degree of disease may have been present in them.

Examination of the brain revealed the presence of a number of spots of softening, of yellowish color, varying in size from that of a marble to that of a pea. The largest of these spots lay in the upper portion of the white substance of the left parietal lobe, and the tissue round it was reddened.

In the right optic thalmus and left corpus striatum were similar spots of rather smaller size, and the posterior third of the outer division of the nucleus lenticularis on both sides showed evident microscopic, though but slight macroscopic signs of a somewhat ill-defined process of the same kind, in the presence of numerous granular corpuscles such as the larger spots also contained.

The internal capsule was but little if at all involved, and that in its posterior part.

The spinal cord was not removed until the following morning, and, owing no doubt to that cause, the subsequent processes of hardening (by Müller's fluid) and coloring were not so satisfactory as could have been wished, and there was difficulty in making as thin sections as were desired. Still, with the aid of Rutherford's freezing microtome and the methods of hardening in mucilage, specimens were obtained which permitted of careful microscopic study.

Before hardening, cuts were made into the cord at short intervals, *but no departure from the normal appearances could be discovered.* The membranes seemed everywhere healthy. After the specimen had lain for a day in Müller's fluid, the surface of several of the cuts, both in the cervical and lumbar enlargement, was scraped, and fine specimens of ganglion cells obtained. Of these almost all contained more, sometimes much more, pigment than is usually met with, but they presented no appearance that was distinctly pathological. Neither was any abnormal condition of the nerve fibres to be made out.

On examination of the hardened cord one pathological change was everywhere visible, namely, thickening of the walls of the arteries, and there was also, in the cervical region, a very slight increase in the connective tissue of one of the lateral columns, giving rise to a slightly heightened blush in the carmine-stained sections. This change did not extend through the whole length even of the cervical cord, most of the sections showing no trace of it. As it was thought to be of but little significance no attempt was made to define its limits.

The ganglion cells of the anterior cornua were normal both in number and outline. They contained, as stated, more pigment than usual, failed to take up the carmine readily, and in a few of them small vacuoles were found.

No great importance could be attached to the loss of reaction to the carmine in the absence of other signs of disease, since this may well have been due to the same putrefactive changes that prevented hardening, the more so that the other cellular elements of the cord were in the same condition in this respect.

The formation of the small vacuoles may have been due to disease, but it also may have been due to putrefactive changes, and at any rate the number of the cells affected in this way was too small to account for the widespread muscular atrophy.

Of the condition of the *peripheral nerves* and of the muscles I am, unfortunately, unable to speak. A portion of the peroneal nerve and of one of the diseased muscles was reserved for examination, but was accidentally thrown away. To the naked eye the nerve seemed to have undergone no change whatever, either in size, consistency or color.

The symptom which presents the most interest in this unusual case, and for which we might have expected the most readily to find an explanation through the *post-mor-*

tem examination, is the rapid muscular atrophy, associated, as it was, with great diminution or entire loss of electrical reaction of the affected nerves and muscles. Only so far as the pathological changes that were actually discovered may be taken as a guide, the lesion which brought about this result must have had its seat either in the muscles themselves or in the peripheral nerves, and, as a matter of fact, the clinical picture was closely like those presented by most of the cases quoted and reported by Leyden in his recent admirable paper,* or others given in the thesis of Dr. J. Gros,† and, indeed, in the case given at greatest length by Leyden (p. 40), and in one or two of those quoted from others, just such slight changes in the spinal cord were found (formation of small vacuoles, increase of pigmentation) as in this case, and good reasons are offered for thinking that they were secondary and of little moment.

The diagnosis by exclusion speaks likewise with great positiveness either for disseminated neuritis or for a myositis with secondary involvement of the mixed nerves. The idea could not be entertained that the lesions in the cerebral ganglia might account for the pain and the rapid atrophy; the cord was essentially healthy; there remains only the nerves and the muscles themselves to consider. That all the symptoms, even including the pain, might be due to an acute myositis is not to be denied on *à priori* grounds; but we are, perhaps, bound to exhaust, first, the better accepted hypothesis, which favors the neurotic origin of such cases as this. It remains, at the same time, to be said that since, in all the cases quoted, the muscles as well as their nerves were found diseased, it is somewhat a begging of the question to say that the nerves were certainly primarily at fault.

* Ueber Poliomyelitis und Neuritis. *Zeitschr. für Klin. Med.*, Bd. 1, Hf. 3.
† Contrib. à l'histoire des névrites. Paris, 1879.

At any rate, if the disease begins in the nerves it does so usually at their peripheral extremities. Thus, in this case, as in very many of those which have been reported, the severest symptoms affected not all the muscles or surfaces of skin supplied by one great nerve trunk, but the parts furthest removed from the central organs, irrespective of the source of the nerve supply,—the legs below the knee, the arms below the elbow, and of these the feet and the hands more than the forearms and the legs.

As regards the changes of sensibility, this rule was in our case most striking, and the same is true of some of the cases reported by others. Thus, in one quoted by Gros* we read:

"24 fevr. Depuis quelques jours, le malade se plaint de douleurs très vives, exclusivements localisées aux deux pieds," * * * etc.

"5 mars. Toujours des mêmes douleurs aux membres inférieurs. Diminution de la sensibilité. Tous ces symptômes restent limités aux pieds et aux régions malléolaires. Depuis deux jours, il accuse des doulcurs de la même nature dans la paume de la main du côté gauche." * * *

In another place (p. 51): "Elle (la sensibilité) est abolie complêtement dans les pieds, dans la région externe des jambes, c'est-à-dire dans le domaine du nerf saphène externe *et dans les branches terminales des autres nerfs.*"

It is not, however, always the case that the peripheral distribution of several nerves is simultaneously and exclusively involved, so much so that Gros lays it down as one of the diagnostic marks of the affection that the symptoms, both sensitive and motor, predominate in the distribution of one or more nerves.

I hope soon to bring further evidence in favor of the opinion which I expressed some years ago, that whether we are dealing with diseases of muscles, nerves or (motor)

* *Loc. cit.*, p. 61.

nerve nuclei, the types of disease are apt to be the same, showing a greater vulnerability on the part of certain muscles, their corresponding nerve fibres and their corresponding nerve nuclei, than is shown by their fellows. It is my belief that this may hold good of the sensitive tracts as well, and that this general law will render clear the recurrence of certain types of diseases involving alterations of sensibility and muscular nutrition better than any simply topographical explanation.

The admission of disseminated neuritis into our nosologies opens a wide field for study, the limits of which are well defined in the monographs to which I have referred. The differential diagnosis has to take account, not only of the so-called ascending spinal paralysis of Landry, of poliomyelitis, perhaps progressive muscular atrophy (Leyden) and lead paralysis of rapid onset (of which I have recently seen a striking illustration), but even of meningitis, as is pointed out by Gros (p. 53).

It is possible that the following case will prove to be an illustration of this fact:

A patient died last summer at the Massachusetts General Hospital, under the care of Dr. B. S. Shaw, who kindly allowed me to examine the case and the specimens, who, throughout her sickness, showed marked symptoms of acute meningitis, viz., extreme pain in the neck, the back and the limbs, greatly increased on even the slightest movement, fever and muscular paralysis, which improved after a time with marked wasting, especially in certain groups of muscles. At the autopsy no sign of meningitis was present.

The cord has not yet been examined microscopically, but seemed to contain minute scattered foci of inflammation.

The pain, which was the marked feature of the whole case, could hardly be explained by the trifling disseminated myelitis, if this proves to be present, while the latter could well have been secondary to an acute neuritis.